SYRINGOCYSTADENOMA PAPILLIFERUM (DEEP SEATED HORIOSTOMATOUS): A CASE REPORT

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ABSTRACT

Syringocystadenoma papilliferum (SCAP) is a rare benign adenexal tumor that frequently shows apocrine differentiation. SCAP usually occurs in the head and neck region in the children. Here in we report a case of 22 years female with SCAP occurring in deeper portion of the neck. The clinical, histopathological features and differential diagnosis of SCAP are also described.

KEYWORDS: Neck, Syringocystadenoma papilliferum, Adenexal

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INTRODUCTION

Syringocystadenoma papilliferum (SCAP) is a rare benign hamartomatous adenexal tumor. Fifty percentof cases are present at birth or in early childhood, while another 15% - 30% case develops during puberty.[1] It typically involves the skin of head and neck. It can be associated with preexisting organoid nevus, sebaceous and other adenexal tumors. Clinical presentation varies widely but the histologic appearance is uniform and characteristic forming the basis of diagnosis.[2] We here present a case of syringocystadenoma papilliferum in deeper portion of neck in a 22 year female.

CASE REPORT

22 year female presented with complain of swelling in the right side of neck region for two years. Initially patient has lesion of 0.5cm x 0.5cm which gradually progressed and was 4cm x 3cm at the time of presentation. Swelling was firm, mobile, non tender. A presumptive diagnosis of cervical lymphadenopathy was made.

The patient was operated under local anaesthesia. Lesion was found just beneath the sternocleidomastoid muscle. Lesion was excised completely with a normal margin of 0.5 cm. Post-operative period was uneventful.

Histopathological Examination:

Gross finding: single mass of tissue measuring 2.2 cm x 0.7cm, black in colour, soft in consistency, cut surface show hemorrhagic area.

Microscopic findings: section showed features of sweat gland tumor with cystic spaces and papillary projections with areas of hemorrhage (Fig. 1). Papillary strands are lined by cuboidal to flattened epithelium forming complex papillary structure at places. The papillary core show dilated capillaries and contain dense lymphoid aggregates and plasma cells with areas showing apocrine metaplasia, focal dysplasia along with lymphocytes and macrophages within the connective tissue (Fig.1 and 2)

The final histopathological diagnosis was syringocystadenoma papilliferum.

DISCUSSION

Syringocystadenoma papilliferum is an exceedingly rare hamartomatous proliferative malformation derived from apocrine sweat glands of the skin. It usually appears at birth or during infancy or around puberty.[1] In this case it started appearing at the age of 22 years.

Majority of SCAP arise on the head and neck.[3] Other unusual reported location of SCAP includeseyelid, buttock, vulva and scrotum, outer ear canal, post-operative scar, thigh, axilla and back.[4]

Early lesion appears as pink or skin coloured solitary nodules, or as grouped nodules. Verrucous and hyperkeratotic lesions can also occur.[5] The lesion enlarge gradually, accelerated growth within a matter of month has also been described.

One third of all the cases arise in an organoid nevus, such as nevus sebaceous.[3,6] Coexisting basal cell carcinoma is noted in 10% of cases.[6] Syringocystadenocarcinoma papilliferum is the malignant counterpart characterized by solid areas and cytologically malignant cells.[1]

Figure 1: Shows numerous papillary infolding lined by cuboidal cells with cystic spaces epithelium and fibrovascular core with lymphocytes

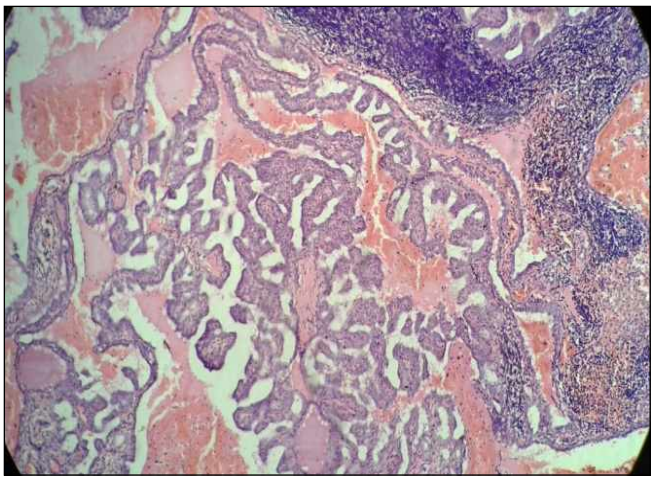
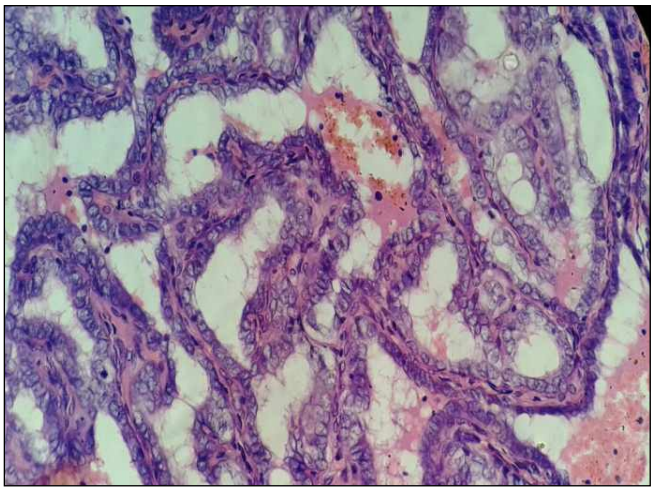


Figure 2: High power view shows papillary projections lined by low to flat cuboidal



These papillary projections and lower portion of the invagination are lined by glandular epithelium typically consisting of two rows of cells.[7] The inner luminal row consists of columnar cells and sometimes shows active decapitation secretion.[8] One significant and diagnostic feature is the presence of a cellular infiltrate mainly composed of plasma cells within the stroma of this tumor particularly in the papillary projections with dilated capillaries.[5,6,10]

Although neck was the common site, in this case lesion was deeper to the sternocleidomastoid muscle which was the unusual site for SCAP. SCAP was not considered as a clinical diagnosis and was a histological surprise. Cervical lymphadenopathy was considered first as provisional diagnosis.

There were few studies which reported the case of SCAP in leg[3], lower abdomen[4], thigh[6] and our case was also at the unusual site, sternocleidomastoid muscle. Surgical excision with reconstruction is the treatment of choice for SCAP.[1, 11] Carbondioxide laser excision of SCAP of the head and neck is the clinical treatment option in anatomic areas unfavourable to excision and grafting.[2,3] Radiotherapy and other destructive procedure are ineffective and best avoided.[11]

In conclusion, SCAP is rare neoplasm with apocrine differentiation. In the present case, lesion was diagnosed as SCAP after histopathological examination with foci of dysplasia which was a rarity. Solitary lesions although in usual location must be excised and sent for histopathological examination to avoid malignant transformation.

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